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MYXOMA-LIKE GROWTHS IN THE HEART, DUE TO LOCALIZATIONS OF SPIROCHAETA PALLIDA*

PLATES 1 TO 4

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In 5 cases of congenital syphilis of infants round translucent nodules have been found in the myocardium, appearing to the naked eye as myxoma-like growths. Three cases were new-born infants dying a few days after birth, one was a child 14 months old, and one a child 18 months old. Of these cases 4 presented signs of syphilis and showed the presence of spirochetes in other organs and tissues; while 1 showed syphilis only in the myocardium.

The myxoma-like nodules occurred in all cases in the anterior wall of the left ventricle, either near the apex or near the ventricular septum. In 1 case a similar nodule was found on the posterior wall of the left ventricle. In 3 cases there were two distinct nodules in the anterior wall of the left ventricle, one just above the apex, the other higher, over the septum. The nodules, which appeared to be nearly round or ovoidal, spherical on section, were all elevated above the general level of the epicardium. The largest were 1 cm. in diameter; the smallest was 5 mm. They extended entirely through the heart-wall from epicardium to endocardium, projecting inwardly as well as outwardly on the epicardial surface. They were light-grayish in color, of a translucent and jelly-like quality, with a few prominent, tho small, blood-vessels running over or around them. Their consistence was that of mucoid tissue. In all respects they resembled small myxomas, and the first ones seen were at first sight so regarded. The hearts of the five infants were much enlarged, dilated, and hypertrophic, the walls of the left ventricle being 2 or 3 times thicker than normal. Smaller translucent areas also occurred throughout the myocardium.

On microscopic examination these round translucent areas were found to consist of an edematous gelatinous connective tissue, showing remains of atrophic muscle fibers, especially at the periphery. Microscopically the nodules were distinctly intramural, not quite reaching the

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endocardium, being separated from it by a thin band of heart muscle. A similar intervening thinner band was also present on the epicardial side. In the center of some nodules the muscle had completely disappeared. The remaining muscle fibers were arrayed somewhat like the meridians of longitude on a globe, farther apart at the equator, and converging at the poles. The space between the pressed-apart fibers was filled in with the mucoid connective tissue. Mucin stains, particularly cresyl violet, gave a strong mucin reaction. The gelatinous tissue was not very cellular, except in areas, the cells being chiefly varied forms of fibroblasts (epithelioid cells), lymphocytes, and plasma cells. Many of the fibroblasts had numerous branching processes, but the branching was not so marked as that in ordinary gelatinous tissue. The intercellular substance was semifluid or mucoid, or consisted of delicate fibrillae in a semifluid matrix. The histologic changes were precisely the same in kind as those between the heart-muscle fibers in spirochetosis of the myocardium, tho differing in degree, in greater mucin content, sharper localization, and greater destruction of muscle fibers. The changes in the muscle fibers indicated a process that was spreading peripherally from a center of more marked change. This central, presumably older, area showed no signs of caseation, fatty change, or fibrosis; it appeared simply more semifluid, or mucoid, than the peripheral portions. No new formation of blood vessels was shown. The cellular infiltration and proliferation were so slight that microscopically the nodule might easily have been taken for an edematous or hydropic area, or, when mucin stains were used, for a myxomatous area in the myocardium.

In sections prepared according to Levaditi's method, these mucoid areas were found to be localized colonies of Spirochaeta pallida occurring in enormous numbers. The smaller mucoid patches throughout the myocardium likewise showed Spirochaeta pallida in great numbers. The larger round sharply localized mucoid areas might be regarded as representing a mucoid granuloma or myxogumma, altho there was no histologic resemblance to the ordinary gumma of late syphilis.

In the literature I have been unable to find any microscopic description exactly fitting these lesions. In the older literature on syphilis, there are not infrequent macroscopic descriptions of the gummata of late acquired syphilis as being semitranslucent, gelatinous, or mucoid in character, but the microscopic picture of such macroscopic gelatinous gummata as described does not in any way resemble these lesions.

Only one instance occurs in the literature of an observation of a syphilitic lesion that was most probably of the same character as those found in my cases.

Shattuck[1] exhibited before the Pathological Society of London, in 1881, a specimen of a "mucous tumor of heart (syphilitic gumma?)," with the following description:

> "The heart of an infant, in connection with the right ventricle of which there had been formed a small circumscribed, slightly nodular and granulated ovoidal tumor, about a centimeter in its chief diameter, which slightly overhangs its base and projects into the ventricle from the anterior wall immediately below the corresponding cusp of the pulmonary valve, the inferior side of which it also involves.
>
> "*Histology.*—A net-work of delicate connective-tissue in the meshes of which lie large many-branching, granular, indented cells, of which the processes form a secondary, closer reticulum. The other organs were healthy. From a child congenitally syphilitic."

Shattuck's observation was made long before the spirochete became the positive identification of a syphilitic lesion, yet his gross and histologic descriptions make it extremely likely that the lesion he described is of the same nature as the mucoid nodules in my five cases, in which Spirochaeta pallida occurred in great numbers, proving conclusively that the lesions were due to syphilis.

If myxoma-like nodules can be produced in the heart-wall by local colonization of spirochetes, the question at once arises as to the true nature of the mucoid nodules described under the heading of myxomas of the heart-wall. The myxoma is regarded as one of the common forms of the relatively very rare primary neoplasms of the heart; but the real nature of these neoplasms has occasioned much discussion among pathologists. I have found reports of 48 such cardiac myxomas in the literature. Of these, 24 were recorded found in the left auricle, 3 in the left ventricle, 2 in the right auricle, 2 in the right ventricle, 1 involving both auricles, 3 on the aortic valves, 2 on the mitral, 8 on the tricuspid, and 3 on the pulmonary valves. The majority were endocardial in origin, either firmly connected with the endocardium, or rising therefrom by a pedicle, giving the mucoid nodule the appearance of a polypoid myxoma. Only in a few cases has the apparent myxoma been imbedded in the heart-wall or shown such intimate relationship as to convey the impression that the growth was intramural, not endocardial, in origin.

These so-called myxomas of the heart have been small in size, and clinically unrecognizable, being discovered only by accident at autopsy. They have been called myxomas because of their translucency and their histologic appearance of branching cells, with fine fibrillae and semifluid intercellular substance. Staining tests for mucin have either not been tried, or have resulted negatively, with the exception of Hanser's[2] case in which positive mucin tests with mucicarmin and thionin were obtained. These reactions Hanser used as positive argument to prove the true myxoma nature of the little polypoid translucent nodule, the size of a cherry stone, which he had found springing from the anterior papillary muscle of the mitral. The location of the growth, the mucin reaction, the absence of blood pigment, and the presence of elastic fibers he regarded as proof of the true neoplastic character of this tumor.

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[1] Tr. Path. Soc. London, 1881, 32, p. 77.

On the other hand, it is asserted that the majority of these endocardial mucoid nodules represent organized thrombi, or inflammatory proliferations of the endocardium, those on the valves in part belonging to the so-called "Lambl's excrescences." I myself have had 2 cases of endocardial mucoid nodules undoubtedly originating as organizing thrombi.

Case 1.—Old acquired syphilis. Angina pectoris. Sudden death. Chronic syphilitic myocarditis. The right auricular appendix was completely filled with a translucent mucoid tissue, attached to the surface of which the remains of an old thrombus could still be seen with the naked eye. Microscopically, the mucoid area presented the appearance of myxomatous tissue, giving good reactions for mucin, mostly devoid of any blood pigment or fibrin, and containing new-formed elastic fibers. Between the myxomatous connective tissue and the thrombus lay an intermediate area of beginning organization and formation of a young gelatinous fibroblastic tissue. Here there could be not the slightest doubt that the myxomatous tissue represented simply the organization of a thrombus attached to the endocardium, for all stages of the process of the organization and development of the mucoid tissue could be seen. As this observation took place before the discovery of the spirochete no examination for spirochetes was made.

Case 2.—Old acquired syphilis. Angina pectoris. Sudden death. Gummatous myocarditis of left ventricle. Thrombosis of left ventricle. Organizing thrombus attached to thinned heart wall just above apex. Next to the heart wall, which at this point was mostly fibroid, the organizing thrombus presented a layer of mucoid tissue, giving reactions for mucin, in chief part containing no blood pigment, and showing new-formed elastic fibrillae. No examination for spirochetes was made.

There can be no doubt that endocardial nodules of mucoid tissue arise from the organization of parietal thrombi, and it is most probable that all the reported cases of cardiac myxoma in which the supposed neoplasm was attached to the endocardium, were not neoplasms, but polypoid formations of a young mucoid fibroblastic tissue, arising from the organization of a thrombus.

[2] Frankfurter Ztschr. f. Path., 1911-12, 9, p. 362.

Fibroblastic proliferations of the endocardium, particularly of the valvular endocardium, show a particular tendency toward the formation of a mucoid connective tissue that gives a more or less pronounced staining reaction for mucin. It has been my experience that in every organizing valvular vegetation, some of the new-formed connective tissue gives in its earlier stages such mucin-staining reactions. The occurrence of mucin in organizing cardiac thrombi has been noted by other writers. Wegelin[3] states that "the mucin-reactions often give a positive result in the organization of thrombi of the heart wall." Brenner, Czapek, Curtis, Lubarsch, and others, also recognize the mucoid character of organizing thrombi.

In not a single one of the reported cases of supposed myxoma attached to the endocardium is the blastoma nature of the mucoid tissue conclusively demonstrated. On the contrary, the evidence given points to the origin from organizing thrombi. They should therefore be rejected as true cardiac neoplasms, and no longer classed with such. A true myxoblastoma of the endocardium is yet to be demonstrated. This applies also to the relatively few intramural mucoid nodules reported as myxomas. From the given descriptions I do not believe that any of them has been a true blastoma. My own five cases show conclusively that mucoid nodules, giving positive mucin tests, occur in the myocardium in cases of congenital syphilis and represent formations of a mucoid fibroblastic tissue due to the localization of large colonies of Spirochaeta pallida. The few intramyocardial myxomas reported (Czapek, Hlava) were probably of the same nature as the nodules which I have described, but Shattuck's case is the only one in which a syphilitic origin was suspected, and the only one in which we may be reasonably sure that the pathologic conditions were the same as in my cases.

I have shown elsewhere that in congenital myocardial syphilis the new-formed interstitial connective tissue is frequently mucoid or edematous in type, and gives a mucin reaction with cresyl violet. In the average case these mucoid proliferations are too small to be seen individually with the naked eye, but may be so extensive as to make the entire heart wall more translucent and lighter in color, as shown in Fig. 2. Larger, more sharply localized colonies of spirochetes rarely produce the larger localized mucoid nodule seen in my cases. These myxogummas are probably relatively rare, or they could hardly have escaped observation to the degree that they have; yet I believe that more detailed examinations of the hearts of congenitally syphilitic infants and children will show a more common occurrence than we now suppose. In my own experience, the myxogumma of congenital syphilis is more frequent than the caseating gumma of the heart in acquired syphilis, of which I have seen only one example.

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[3] Frankfurter Ztschr. f. Path., 1911-12, 9, p. 117.

SUMMARY

Localized colonization of Spirochaeta pallida in the myocardium in congenital syphilis may lead to the formation of an intramural mucoid nodule, from 5 to 10 mm. in diameter, resembling mucoid tissue both macroscopically and microscopically, and giving a positive mucin reaction. Such myxogummas are relatively rare, and may be mistaken for myxomas.

A mucoid fibroblastic tissue giving positive mucin reactions is very commonly produced in the organization of parietal cardiac thrombi and valvular vegetations. The absence of blood pigment, the positive mucin reaction, and the presence of newly formed elastic fibers cannot, however, be taken as points deciding the blastoma character of the mucoid tissue.

There is no positive proof of the occurrence of a true myxoblastoma of the heart. The mucoid nodules attached to the endocardium are undoubtedly organized thrombi; the rarer ones in the myocardium may most plausibly be interpreted as myxogummas, or proliferations of a mucoid fibroblastic tissue caused by the localization therein of Spirochaeta pallida.

EXPLANATION OF PLATES

PLATE 1

FIG. 1. Heart of infant dying from congenital syphilis. Two mucoid nodules (myxogummas) in anterior wall of left ventricle near septum. Left ventricle hypertrophic and dilated.

FIG. 2. Heart of infant dying from congenital syphilis. Two mucoid nodules in anterior wall of left ventricle, above apex and involving part of septum. Entire heart light-colored and more translucent than normal as a result of diffuse interstitial mucoid proliferation or edema caused by diffuse spirochetosis. Mucoid nodules contained enormous numbers of spirochetes.

PLATE 2

FIG. 3. Transverse section of mucoid nodule seen in Fig. 1. Nodule extends through myocardium, is elevated on epicardial and endocardial sides, but separated from epicardium and endocardium by narrow bands of muscle. The bluish fibrillar areas in the nodule are the mucoid areas containing spirochetes. Bands of pale atrophic muscle found throughout entire translucent area. The appearances are those of localized myxedema of the myocardium with atrophy and degeneration of muscle rather than of localized granulomatous proliferation (gumma). Hematoxylin and eosin.

PLATE 3

FIG. 4. Section of translucent area stained with cresyl violet. Pink fibrillae show marked mucin reaction between muscle fibers.

PLATE 4

FIG. 5. Section of the myxoma-like nodule in Fig. 2, from half of nodule prepared by Levaditi's method. Drawing made from photomicrograph showing actual number of spirochetes appearing in one focal field, with those partly out of focus represented as in one focal plane.

PLATE 1

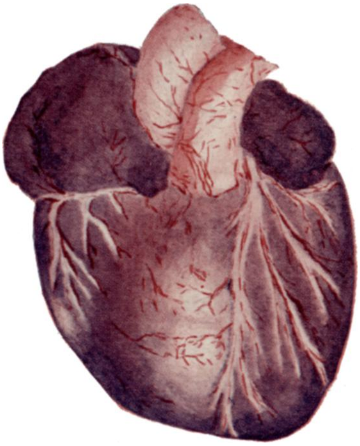

Figure 1

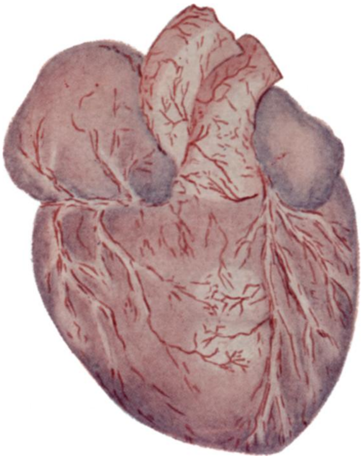

Figure 2

PLATE 2

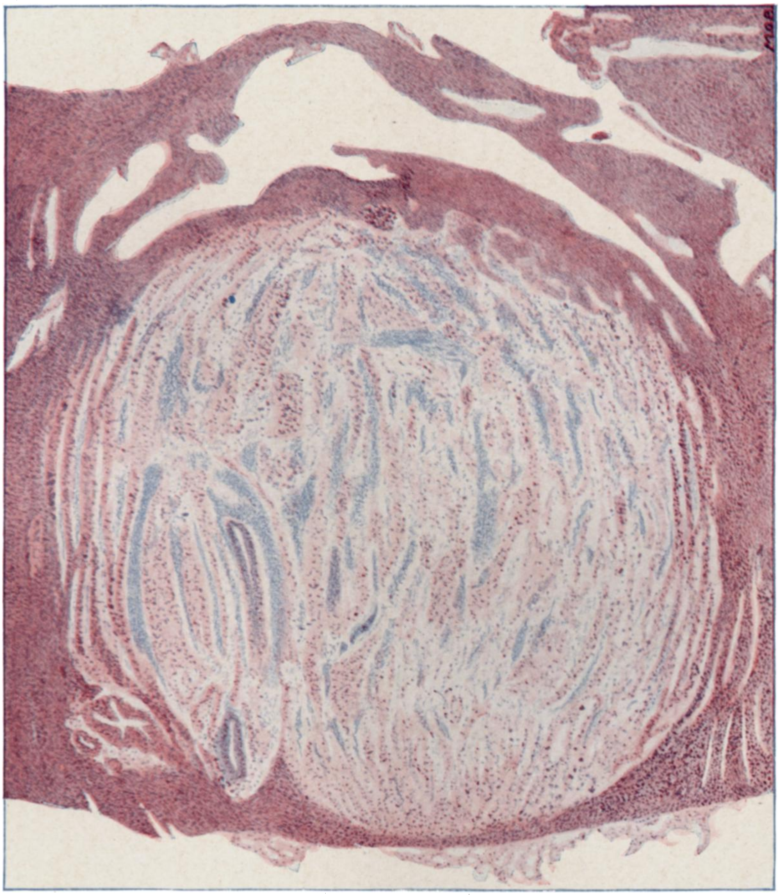

Figure 3

PLATE 3

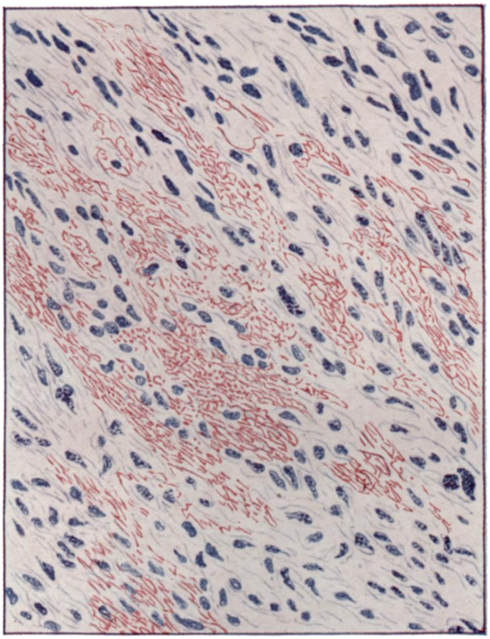

Figure 4

PLATE 4

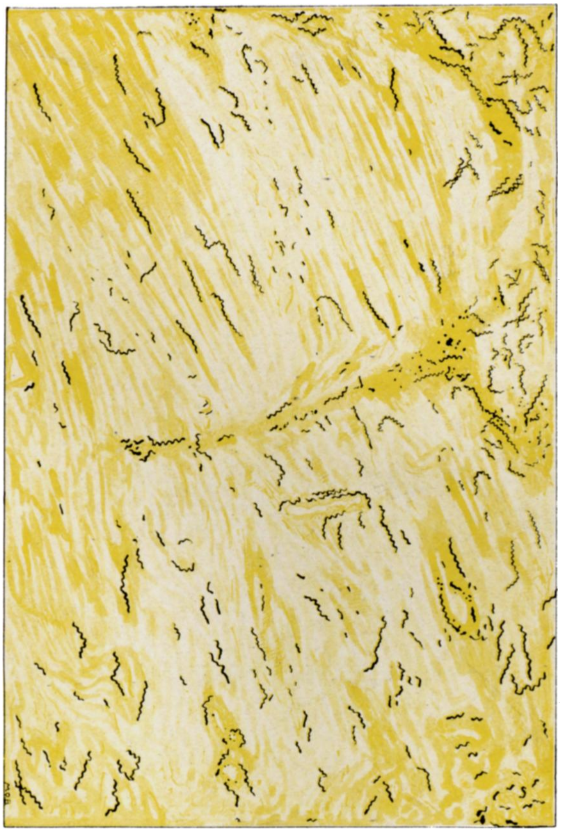

Figure 5